Case of rheumatic heart disease in medicolegal autopsy

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Abstract
Rheumatic heart disease (RHD) still remains one of the leading causes of death in children and young adults. It is a major public health problem owing to the issues of overcrowding, poor hygiene and low adherence to secondary prophylaxis. We present a case report of adolescent orphanage girl died in suspicious manner and found to have RHD on histopathological study of her heart. This case highlights importance of early diagnosis of the disease that would have possibly avoided preventable cause of death in teenage girl. The interesting findings we noticed in this case report is presence of myxoid area of left ventricle which showed extensive collection of Anitschkow cells and chronic inflammatory cells, and we didn't get reference of similar findings in the literature search of rheumatic heart disease. The heart with pericardium together weighted 620 gm which is maximum weigh we encountered in our experience.

Keywords: RHD, Anitschkow Cells, Aschoff bodies, Autopsy.

Introduction
Rheumatic heart disease (RHD) follows the severe acute or recurrent rheumatic fever. It remains a major public health problem in India and also in several developing countries.[1] It becomes the leading cause of death in children and young adults, if untreated. Currently an estimated 33.4 million people are living with RHD worldwide.[1] RHD remains a major public health problem owing to the issues of overcrowding, poor hygiene and low adherence to secondary prophylaxis.[2] Here, we present a case report of adolescent orphanage girl died in suspicious manner and found to have RHD on histopathological study of her heart. This case highlights importance of early diagnosis of the disease that would have possibly avoided preventable cause of death in teenage girl.

Case Report
A 13 year old orphanage girl was brought to casualty stating that she is in unconscious state since one hour duration. On examination, she was found dead and confirmed as "brought dead". She was brought to casualty by the caretaker of orphanage centre located in outskirts of the city. There was past history of jaundice one month back, for which she took native Ayurvedic treatment, and not consulted any medical professional nor visited any hospital. Past history revealed illness like recurrent sore throat infections, fever and joint pains, as revealed later by her associates in orphanage. There was no history of proper medical treatment taken for those complaints. It was also found that, there was not much hygiene maintained in that orphanage centre and there was no medical consultant attached to it, in case of any illness to the inmates. Police suspected a foul play by caretaker and initiated postmortem examination in view of unnatural death in young teenage girl. A medicolegal autopsy was performed on her to know the exact cause of death after obtaining permission from local magistrate. During postmortem, vaginal swabs were also sent for sperms, which were 'negative' for spermatozoa.

Gross Findings
On opening the chest cavity, the pericardial cavity was found to be filled with inflammatory exudate amounting to 200 ml. The heart appeared massively enlarged and together with pericardium it weighted 620 gm and after stripping pericardium it weighted 461 gm (Fig. 1). The pericardium was tightly adherent to the underlying heart due to presence of thick fibrinous exudate resulting in classical "bread and butter" appearance. The pericardium was grossly thickened and fibrotic. Further cut sections of pericardium showed granular fibrinous exudate seen attached to the visceral surface of the heart. There were multiple petechiae spots noticed over the posterior surface of heart. Heart was cut opened by Virchow's method (along the flow of blood). On opening the heart, the right atrium was dilated and right ventricular wall thickness was 0.4cm. The Mac Column plaque was identified in the left atrium which was gray white and fibrotic, thickened area (Fig. 2). The left ventricle showed asymmetric wall hypertrophy with thickness of 2.1cm near the atrio-venticular (AV) region. Interestingly there was patchy grey white to myxoid areas seen near AV region measuring 1.5x1 cm (Fig. 2-blue circle). Interventricular septum (IVS) showed asymmetric thickness of 0.8cm in the middle. Mitral valve was thickened and fibrotic and its edges showed granular excrescences on atrial surface and patchy grey-white areas on ventricular surface. Several bits were given from representative areas.

Microscopy findings
Histopathological examination from pericardium showed extensive fibrinous exudate with edema and mixed inflammatory infiltrate suggestive of "fibrinous pericarditis" of rheumatic heart disease (Figure no 3A). The pericardium also showed extensive fibrosis and the inflammation was extending into pericardial adipose tissue. Section from left atrium showed subendocardial fibrosis with granulation tissue formation surrounded by lymphocytes and plasma cells with features consistent with "Mac Callum plaque"

(Fig. 3B). Section from left ventricle showed "Myocarditis" with formation of Aschoff bodies composed of "Anitschkow cells" displaying elongated Caterpillar shaped nucleus, and surrounded by lymphocytes and plasma cells (Fig. 4 A,B,C). Interestingly the myxoid area of the left ventricle showed extensive collection of Aschoff bodies containing Anitschkow cells scattered in singles and small aggregates admixed lymphocytes and plasma cells extending from subendocardial area to pericardial surface transmurally. The nucleus of Anitschkow cells showed elongated "caterpillar" appearance when cut horizontally and "Owl eye" appearance with perinucleolar clear halo when the section is cut vertically (Fig. 5). The sections from musculature of left ventricle and Interventricular septum showed enlargement of nucleus suggesting hypertrophic changes.

In the same case, the sections from the lung revealed features of chronic venous congestion and pulmonary edema. Sections from the liver also showed chronic venous congestion. Sections from the kidneys and spleen were unremarkable. In view of classical findings of Pancarditis, the final diagnosis of Chronic Rheumatic Heart Disease was issued.

Fig. 1: Gross specimen of Heart: Showing thick adherent fibrous pericardium (A) displaying "bread and butter" appearance; and massive enlargement weighing 461gm (B).

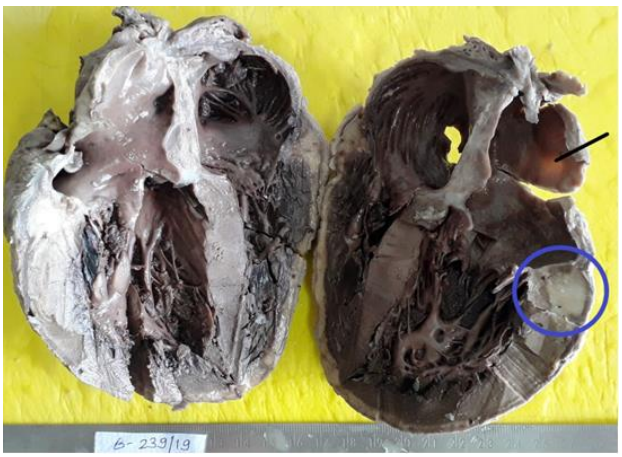

Fig. 2: The Mac Column plaque was identified in the left atrium (black arrow) which was gray white and fibrotic, thickened area; with patchy myxoid area (blue circle) in left ventricle near AV region.

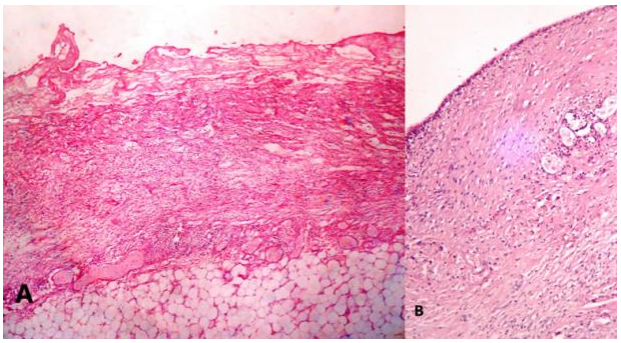

Fig. 3: Pericardium showing extensive fibrinous exudate with inflammatory cell infiltrates suggesting fibrinous pericarditis **(A).** section from left atrium showing subendocardial fibrosis with granulation tissue formation consistent with "Mac Callum plaque" **(B).** (H & E stain, 4x magnification)

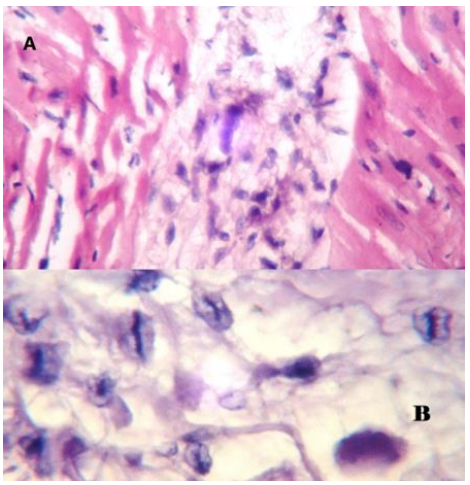

Fig. 4: Section from left ventricle showing Myocarditis with formation of Aschoff bodies/ Rheumatic granuloma **(A).** composed of "Anitschkow cells" displaying elongated Caterpillar shaped nucleus **(B).** (H & E stain, 20x and 40 x magnification respectively)

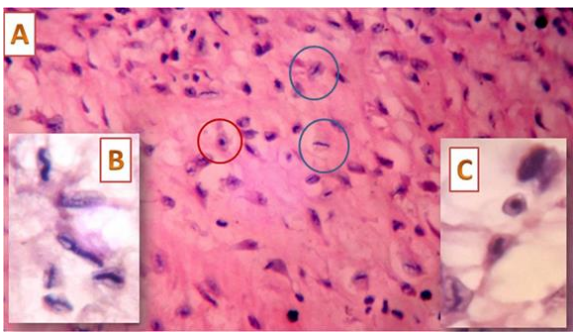

Fig. 5: Myxoid area of the left ventricle showed extensive collection of Aschoff bodies containing Anitschkow cells. **(A)** the nucleus of which is elongated "caterpillar" appearance when cut horizontally. **(B** and blue circles in A) and "Owl eye" appearance. **(C** and red circle in A) when cut vertically. (H & E stain, A is 10x; B & C are 40 x magnifications)

Discussion

Rheumatic heart disease (RHD) is the cardiac manifestation of rheumatic fever (RF). RF is an autoimmune response to infection caused by streptococci usually diagnosed as acute sore throat. Around 3-5% of cases develop RF following the group A beta hemolytic streptococcal infection.[3] As the acute illness resolves, the valvular lesions persist and tends to progress over time to chronic RHD.[4] Rheumatic carditis is the active inflammation of the heart tissues, most importantly the mitral and/or the aortic valves, caused by acute rheumatic fever. Rheumatic heart disease (RHD) is a chronic valvular disease caused by heart valve damage from severe or repetitive episodes of acute rheumatic fever (ARF).[1] The disease is more common and severe in females than in males. Although the disease rarely occurs, it has the mortality rate of 2–5%.[5] The Histologic Hallmark of Acute Rheumatic carditis is "Aschoff bodies" and three stages are described in its development. Early or exudative stage is characterized by swelling and edema of the collagen (fibrinoid degeneration). Intermediate or proliferative/granulomatous stage shows infiltration by lymphocytes, histiocytes, fibroblasts, and Anitschkow cells. The prominent palisading by histiocytes around the fibrinoid area gives it a granuloma-like appearance (Hence called as Rheumatic Granuloma). This phase lasts up to the 13th week of disease. Following which healing of the lesion occurs, the late/senescent phase.[6] Once the heart is affected by a myocarditis, then it is prone to get reactivation and recurrence of the disease.[7]

The interesting finding we noticed in this case report was massive hypertrophy of cardiac musculature especially in the left ventricle and Interventricular septum. The myxoid area of left ventricle showed extensive collection of Anitschkow cells and chronic inflammatory cells, and we didn't get reference of similar findings in the literature search of rheumatic heart disease. We had reported similar classical case of RHD with Mac Column plaque formation previously in a female patient which was again a medicolegal case.[3] Hence it is important to prove the exact cause of death by careful examination of every organ sent to pathologist and thereby save the victimized persons from alleged litigations.

As quoted by Negi et al, the exact data on incidence and prevalence of RHD in India is lacking and prevalence estimated in school age group is underestimation of disease burden.[9] It is surprising that in this antibiotic era of 21st century, we still come across RHD especially in lower socioeconomic group due to lack of education and ignorance about the consequences of untreated streptococcal pharyngitis. The RF and RHD represent diseases of poverty, overcrowding, poor sanitation and lack of affordability to quality health care.

One of the largest review of RHD was done by Seckeler et al, in which 164 articles reviewed from 1970 to 2009. They conclude that, recognition of subclinical carditis will identify those children who would benefit from secondary antibiotic prophylaxis and hopefully prevent the progression to clinically significant RHD.[10]

Preventive strategies for RHD are a set of well-established measures aimed at preventing manifestation of the disease or its complications. The World Health Organization (WHO) global action plan targets a 25% reduction in premature mortality from non-communicable diseases by the year 2025. Control and prevention of RHD will play an important role in achieving this goal. Preventive strategies include primordial, primary, secondary, and tertiary prevention, each of which is specific for certain susceptible groups and situations.[8]

Conclusion

India, being one of the fast developing countries, the prevention of infection and prophylaxis with appropriate antibiotics should be necessary to decrease the burden of disease. Primary care physician diagnosis and follow up of the case will definitely play important role in decreasing the mortality and complications of the disease. This medicolegal autopsy case demonstrates the classic histopathology features of RHD, reveals the importance of careful examination of autopsy findings during postmortem and thorough histopathological examination of organs sent.

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Conflict of interest

None.

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